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Clinical Cases.

CASE IN WHICH ATTACKS OF INTERMITTENT TONIC MUSCULAR SPASMS, IMMEDIATELY FOLLOWED BY COMPLETE TEMPORARY PARALYSIS, HAVE FREQUENTLY AND PERIODICALLY OCCURRED DURING THE ENTIRE LIFE OF THE PATIENT, THE HEALTH IN THE INTERVALS BEING NORMAL.

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Antecedent History.—The patient is a young woman aged 17. Her family history is unimportant.. The mother states that during utero-gestation she herself suffered from nervous symptoms as the result of fright, from being chased by a mad dog. She nursed the child for a year, but it was always very delicate and undersized, and was unable to walk till it was three and a half years old.

At birth the patient is stated to have been apparently healthy, but three days afterwards she was seized with a "fit." After this first attack, which was of a convulsive nature, she had no definite illness, but was subject to frequent "faintings," in which she lay for a short time motionless, and "turned up the whites of her eyes." When three years old, it was for the first time observed that in these seizures the limbs were "drawn up." These periodical attacks have continued since, but have become more frequent and severe. Two or three years ago they were accompanied by screaming and other apparently violent hysterical symptoms, but of late these have disappeared. The catamenia have never occurred. In the interval between the attacks the physical health has been good, but mentally the patient is considered deficient, and although she has been to school, she cannot read or write like other girls of her age.

Present condition—A. *During the intervals of the attacks.*—The patient is pale and delicate-looking, and seems several years younger than her age, but all the organs and functions are normal, the body is well nourished, and the general health is good.

She has a peculiar sardonic expression, owing to the lower lip being tightly contracted round the teeth, over which the upper lip projects. There is marked flattening of the skull in the region of the posterior fontanelle. The movements of the eyeballs are somewhat restless and unsteady, and there is difficulty in fixing them on any object, but there is no strabismus or otherwise deficient action. The pupils are equal and normal, and contract to light and accommodation. Vision is natural. The mouth and face are symmetrical, but their movements are accompanied by slight choreic-like twitchings. The tongue is straight, and its actions are apparently normal, with the exception of a slight restlessness on movement. The articulation is not very distinct, but without marked peculiarity. The patient is cheerful, and willingly assists in the work of the ward; she answers questions of an ordinary kind, and conducts herself in a way which has not caused any special comment from the nurses. Her intelligence is, however, evidently defective. She is slow and stupid, childish in her manner, cannot give an account of her illness, has a bad memory, and cannot read or write. The special senses are all intact. The appearance, movements, and strength of the limbs are normal in every respect, except that there is a constant slight restless or choreic-like movement of all the extremities. The grasp of the hands and all the actions of the upper limbs are as in health. Locomotion is normal, and the patient can walk and move about like other persons. There is no rigidity or wasting of the muscles, which have a natural appearance. Sensibility of the skin is everywhere normal. The knee-jerk phenomena are distinctly increased, but the plantar reflexes are normal. The mechanical irritability of the muscles is not markedly augmented. The electrical reactions are somewhat difficult to determine with delicacy, owing to the nervousness of the patient, and to the choreic-like movements of the limbs. They appear to be everywhere normal, with the following modifications. 1st. Stimulating the *muscles* with the galvanic current causes more vigorous contraction with the same strength of current, than when the trunk of the *nerve* which supplies them is acted upon. 2nd. In both muscles and nerves the AOC is slightly but distinctly greater than the ACC; the KCC being greater than either, and KOC is not obtainable by the strongest currents which could be tolerated.

B. *Condition during the attacks.*—This case having been under observation for upwards of a year, the condition during the so-called attacks may be summarised as follows. The patient, while in her usual healthy state, suddenly, and without apparent cause, experiences a peculiar sensation which she cannot describe, but which indicates that a seizure is imminent. She at once goes to her mother, who observes "the veins in her forehead swelling." A few minutes afterwards a sudden tonic spasm of certain muscles takes place, during which they become tense and hard, and which contract the limbs in different directions, so as to cause a variety of distortions. The parts thus affected are immovable and rigid, and their position cannot be changed by voluntary effort or even by considerable active force. When the legs are thus attacked, the patient falls down, owing to the violently produced deformity which ensues. There is no loss of consciousness or intellectual disturbance, and only when the contractions are unusually severe is there pain of a cramp-like character. This condition having lasted for about five minutes, the rigidity rapidly relaxes, and is succeeded by complete paralysis in the same distribution, in which the limbs hang flaccid, useless, and incapable of any voluntary movement. This state continues for about five minutes, after which gradually the muscles regain their power, and in from ten minutes to a quarter of an hour the patient is in a perfectly normal condition. This constitutes one attack. For the past few years the rule has been for these seizures to occur at irregular intervals, many times a day for about a week, after which there is a fortnight of complete freedom, during which time the patient is quite well. Then occurs another week of attacks, and so on, with fairly regular occurrence. These seizures are not all of equal severity. Most commonly one or more limbs only are affected; but occasionally the entire body is in a state of rigidity, every voluntary muscle apparently being in a state of violent contraction. Frequently a single limb only is involved; usually both extremities of the same side; sometimes one arm and the other leg; more rarely the two arms, or the two legs; and occasionally the entire body. When the limbs are the subject of this spasm the effects are not always the same, the muscles do not seem to be always attacked with the same proportionate severity, and consequently the deformity varies, and this may occur during different attacks without apparent definite order. Some of the chief attitudes assumed are portrayed in the annexed drawings, which, however, do not give a complete idea of the intense muscular contraction which deforms the limbs. In the upper extremity there are three prominent types. 1. The upper arm adducted and rotated

inwards and backwards; the forearm extended and pronated, the wrist flexed, the proximal phalanges of the fingers flexed and their two terminal phalanges extended; the thumb extended and abducted; the entire limb projecting in a straight line posteriorly, the palm of the hand looking backwards and upwards (Fig. 1). 2. The upper arm abducted

Fig. 1.

and at right angles to the trunk; the forearm flexed; the wrist, fingers and thumb flexed; the clenched fist resting on the shoulder (Fig. 2). 3. The upper arm adducted, with the elbow close to the side; the forearm flexed; the wrist extended; the proximal phalanges of the fingers and thumb extended; their distal phalanges flexed; the hand resting on the shoulder, the palm upwards (Fig. 3).

In the lower limb there are also three chief types of de-

formity. 1. The thigh completely extended; the leg flexed; the foot extended; the toes flexed; the plantar aspect of the foot looking upwards and forwards. (Fig. 1.) 2. The thigh partly flexed and adducted, so that the knee crosses the other limb; the leg slightly flexed; the foot inverted; the toes flexed. (Fig. 2.) 3. The thigh, leg, foot and toes extended, so that the limb is in a state of rigid straight extension, the toes

Fig. 2.

Fig. 3.

pointing downwards, the foot being in the position of talipes equinus. (Fig. 3.)

Even these three main types of deformity in the limbs undergo various minor modifications. When the entire body is attacked, one or more of the positions described of the limbs may be assumed, and are generally different in the two sides, so that a most irregular appearance is the result. (Fig. 4.) In addition, the muscles of the face are contracted and rigid, and

the countenance distorted in a variety of ways; the eyelids are sometimes forcibly closed, sometimes widely open. The eyeballs generally are fixed upwards and inwards, so that only the white of the eyes can be seen. The neck is stiff and rigid, the head being generally forced backwards till it touches the back. There is evidently obstruction to the breathing, possibly due, partly to contraction of the muscles of the neck, and partly to fixation of those of the thorax. The spine is arched and in a condition of complete opisthotonus. During such an attack the senses are acute; the muscles are all hard and tense, there is severe cramping pain, and considerable dyspnœa. Swallowing is impossible, speech is prevented, and the skin is bathed in profuse perspiration. This is succeeded by complete temporary general paralysis, in which the limbs hang limp and perfectly useless. The nature of any seizure cannot be predicted, and the extent and distribution of the deformities succeed one

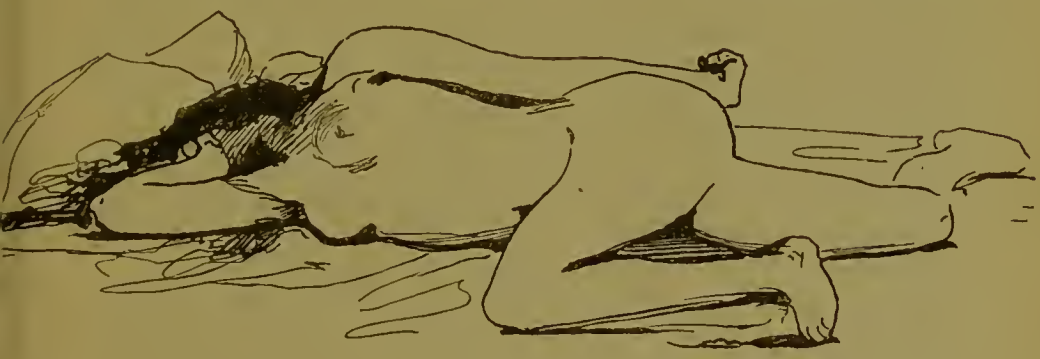


Fig. 4.

another without method or regularity. The precise order in which the individual muscles were attacked could not be determined with any degree of exactitude on a sufficiently extensive scale to form any generalisations, as the physician himself did not frequently observe the entire paroxysm, and the statements of the nurse were not sufficiently accurate. As far, however, as could be ascertained, the most common sequence of events was as follows: Contraction begins in the left hand, spreads upwards to the forearm and arm; then involves the left foot, and subsequently the rest of the lower limb; afterwards the face, eyeballs and head, which last is drawn backwards and towards the left. No exciting cause for the paroxysms could be determined, except that they were liable to be induced by mental emotion and excitement. They never took place during sleep, and were not brought on by voluntary motion or artificial stimulation.

Progress of the Case.—No essential change has taken place

in the last fifteen months, during which the patient has been under observation. The attacks still occur with their former frequency, but of late their severity has somewhat diminished. Treatment of a variety of kinds, including tonics, sedatives, and electricity, has been followed by absolutely negative results. During the spasmodic attack, friction of the affected parts usually gave relief. For the past few months the general health of the patient has been unsatisfactory, and there is a suspicion of incipient phthisis: otherwise her condition is as before.

Commentary.—The clinical picture just portrayed does not exactly correspond with any of those diseases to which a distinguishing name has been given. I have therefore been compelled to describe the case under the appellation of its chief and characteristic symptoms, namely, attacks of intermittent tonic muscular spasm, immediately followed by complete temporary paralysis beginning three days after birth, occurring frequently and with almost regular periodicity throughout the entire life of the patient, the health between the seizures being normal. Although the sequence of events detailed does not in all respects answer to any of the typical descriptions of our text books, it at the same time presents certain features in common with some of them. These it may be well to notice, with the view of determining whether the affection under consideration may not be a modified form of some well-known disorder.

In so-called "Thomsen's disease," for example, there are temporary attacks of cramp which may have existed during the whole life of the patient; but between this and the condition of our patient there is an important distinction to be made. In Thomsen's disease the muscular spasm is directly caused by voluntary efforts, and by them alone. In the case before us the contractions are spontaneous, and in no way affected by active movement.

To the severe forms of Tetany this case bears some resemblance. The intermittent nature of the spasm is common to both. On the other hand, these two affections diverge in many important particulars. The disease under consideration having begun immediately after birth, and continued ever since; the attacks being unaffected by external irritations, and the characteristic hyper-excitability of the nerves on electric stimulation being absent; the temporary but complete paralysis after each seizure, and the impaired mental faculties; all warrant us in removing it from the typical category of Tetany.

To the contractures of hystero-epilepsy this case might at first sight appear to bear a likeness, and the annexed drawings of some of the deformities presented by the patient are some-

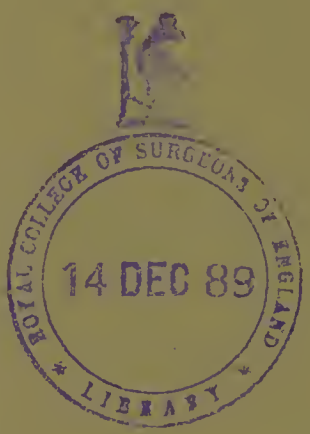
what similar to those delineated in the beautifully artistic representations of M. Richer in his great work.[1] Here, however, consciousness was never lost during the attacks, and there was a total absence of that train of other symptoms characteristic of hysteria. It is true that there was a somewhat vague account of the girl's conduct, lasting for a few months only, which suggested that disorder, but this, if present, was probably a complication of her already existing malady.

This case must therefore stand by itself, as it cannot be placed under the head of any of those affections in which there is a typical clinical history, condition, or progress. Although its leading features have already been detailed, I may now summarise the salient points, those which are unusual, and which render it specially worthy of attention. 1. The disease began at the early age of three days, and has continued without intermission for seventeen years. 2. The most prominent symptoms were attacks of intermittent tonic muscular contractions of varying severity, and involving varying groups of muscles. 3. The seizures of muscular contractions were periodic, and occurred at almost perfectly regular intervals during every third week of the patient's life. 4. The muscular contractions occurred spontaneously without apparent cause, and were not induced by voluntary motion, by friction or other external stimuli, and the electrical excitability of the nerves and muscles was not increased. 5. The muscular contractions were, as a rule, painless. 6. The contractions followed no regular order, and were accompanied by no constant or special deformity, such as the so-called gynecological hand. Most frequently the spasm began in the left fingers, extended from them up the arm to the face and head; from thence to the foot, and up the leg and thigh of the same side. 7. The attacks of spasm were immediately followed by complete but temporary paralysis, which was exactly confined to the same regions, and in which there was no trace of muscular rigidity or voluntary movement. 8. The general health of the patient in the intervals of the attacks was robust throughout, except the slight choreic movements on voluntary motion, and some degree of mental deficiency.

The pathology of this condition must remain for the present a matter of speculation. It is improbable that the muscles themselves are diseased. There is no evidence that the nerves are affected. The seat of lesion must therefore be situated in the nervous centres, probably in the brain, and possibly in the motor areas of its cortex. Its nature we cannot demonstrate. It is scarcely likely that any gross organic degeneration is

[1] 'Etudes Cliniques sur l'Hystéro-Épilepsie ou Grande Hystérie.' Paris, 1881.

present. We must therefore assume the disturbance to be of dynamic origin and due to delicate molecular trophic changes, the existence of which is hypothetical. Hence this disorder may be looked upon as a neurosis. The intermissions, the periodicity, and the paralysis following the spasms, would seem to indicate a continued irritation of ganglionic cells in the grey matter, which causes explosions at intervals, as evidenced by the motor paroxysms, which excess of energy is followed by exhaustion in the shape of paresis. The occasional order in which these phenomena take place is suggestive of this irritation being situated in the cortical substance of the cerebrum.



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